VOL. 36. APRIL, 1909. No. 4.

The Journal
OF
Nervous and Mental Disease

Original Articles

SACRAL TABES: A CASE WITH NECROPSY[1]

By S. Leopold

ASSISTANT IN NEUROPATHOLOGY IN THE UNIVERSITY OF PENNSYLVANIA

(From the Department of Neurology and the Laboratory of Neuropathology in the University of Pennsylvania)

Notwithstanding the fact that clinical cases of tabes, with preservation of the patellar tendon reflexes have been reported, the opportunity rarely arises to study this phenomenon pathologically. As far as I have observed, in a review of the literature two other cases, one by Achard and Levi, in the *Nouvelle Iconographie de la Salpêtrière,* Vol. XI, the other by T. Pineles, in *Obersteiner's Arbeiten,* 1896, have been reported with pathological findings, similar to those in this case. However, Redlich[2] and Pick[3] each have reported cases in which the sacral segments were chiefly involved, but these were cases of tabes dementia paralytica.

The patient, G. W., æt. 54, was admitted to the Philadelphia Hospital and was in the service of Dr. W. G. Spiller, through whose kindness the clinical history and pathological material were obtained.

The clinical history shows that one and one half years before admission the patient commenced to have sharp pains confined to his right thigh. Since his admission he has been confined to bed because of his ataxia. He has had incontinence of urine and feces during this period.

The family history and previous medical history are negative.

Examination shows an ill-nourished, markedly emaciated old man. Right eye shows a cataract, the cornea is ulcerated. Left pupil very myotic and reaction to light is questionable. Ocular movements are normal, associated movements are good. There is emaciation in the muscles of the shoulder girdle and upper limbs. The muscles are flaccid and soft, hypotonicity is distinct in the upper limbs. Power, as shown by the grip test, is fair on the right and slightly more on the left.

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[1] Read before the Philadelphia Neurological Society, November 27, 1908.

Triceps and biceps tendon reflexes are present on each side.

The ataxia as shown by finger-to-nose test is slight. The lower limbs present the same general emaciated condition of the muscles; power and resistance to passive motion are diminished. In heel-to-knee test, ataxia is present in both limbs. There is no tenderness over nerve trunks, and no fibrillary tremors. Achilles tendon reflex is absent on both sides. No Babinski or ankle clonus.

The patellar tendon reflex is obtained on each side, and is very prompt, especially in view of the marked wasting of the thigh muscles. While the leg is not thrown forward on the right side, and only slightly on the left, the contraction of the quadriceps is marked.

No mental disturbance is to be noted; he remembers past events fairly well.

Several months later the patellar tendon reflexes were examined again by Dr. Spiller and found to be of normal response.

At a later date, some difficulty was experienced in obtaining these reflexes, but a prompt response occurred on the left side, as shown by contraction of the quadriceps muscle, with very little movement of the leg. On the right side the reflex was not quite so prompt. At this time the eye examination showed the pupils contracted and reaction to light questionable.

Clinical Summary.—The hypotonicity and ataxia, the shooting pains of the lower limbs, the loss of Achilles tendon reflexes, the vesical and rectal symptoms, the myosis, make it clearly a case of tabes dorsalis. The prominent feature of the clinical history is the nearly normal condition of the patellar tendon reflexes with so pronounced tabetic symptoms.

Sections were examined from the following segments of the spinal cord: The eighth cervical, eighth thoracic, first, second, third and fourth lumbar. From the fifth lumbar the cord was cut into small pieces, as it was impossible to divide it according to the root segments. All the sections were stained with the Weigert, hemalum and Nissl stains.

From the last sacral segment to the fifth lumbar the posterior columns were markedly degenerated, especially on the right side, involving the whole of the root entrance or cornu radicular zone of Marie, which contains the medium and long fibers, forming the reflex collaterals and ascending fibers of the posterior columns. On the left side the root entrance zone was much less

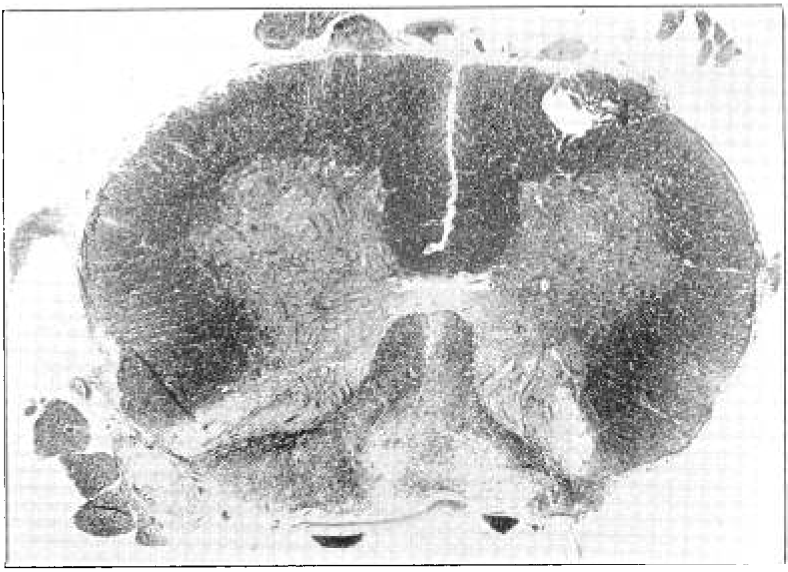

Fig. 1. Photograph of a section from the fourth lumbar segment. The hole in the anterior column indicates the right side.

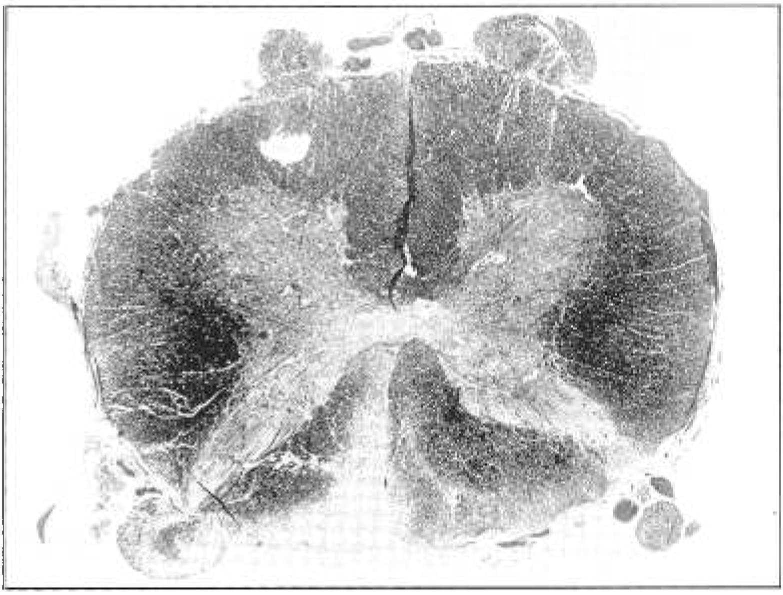

Fig. 2. Photograph of a section from the third lumbar segment. The hole in the anterior column indicates the right side.

destroyed and many normal fibers entered the posterior horn. The pathological picture corresponded to the clinical findings of vesical and rectal disturbance, and loss of Achilles tendon reflex. In view of the preservation of some of the reflex collateral fibers on the left side one might have expected some evidence of the Achilles reflex on this side. However, we know that it is not necessary for all the fibers to be destroyed to cause the failure of the reflex. This is well proven in those cases of tabes with secondary hemiplegia in which a return of knee-jerk occurred.

Fourth Lumbar Segment.—At this level the root entrance zones are markedly degenerated, the sclerosis extends to the posterior horns. The posterior roots are very much degenerated, while the anterior roots are normal.

Third Lumbar Segment.—In this segment the sclerosis is wedge-shape, with its base at the periphery, the apex at the posterior commissure. This triangular area of degeneration is more marked on the right side, its extent being less than in the fourth lumbar. This is the highest level at which the posterior root fibers entering the cord are distinctly degenerated. The sclerosis of the posterior roots of this segment is slight and nearly equal on the two sides, though the right side is a little more degenerated. The anterior root fibers are normal. The pathologic findings, in this segment which shows the nearly complete preservation of the reflex collaterals, explain the clinical phenomenon of the preservation of the patellar tendon reflex.

Second Lumbar Segment.—The triangular area of sclerosis on the right side is smaller than in the preceding segment. A corresponding degree of change is noted on the left side, the area being less pronounced. The root entrance zone on the right side is considerably sclerosed along the periphery of the cord, although the fibers entering at this level seem to be intact. This degeneration is probably due to the loss of ascending fibers from lower segments. On the left side the root entrance zone is about normal. The posterior root fibers show some slight sclerosis. The anterior root fibers are normal.

First Lumbar Segment.—In this segment the root entrance zone on the left side is normal, the right root entrance zone shows very slight sclerosis. The triangular area on the left being faintly noted as similar to the right side. The left posterior root fibers are almost normal, the alteration is so slight as to be questionable, the right posterior root fibers show very slight degeneration. The anterior root fibers are normal. In this section as well as in the second and third lumbar segments, the root fibers were cut from the cord and many sections studied.

Eighth Thoracic Segment.—The column of Goll on the right side is much more degenerated than on the left. The columns of Burdach, root entrance zones, and cornu-commissural zones on each side are not involved. The anterior and posterior root fibers show no changes.

Eighth Cervical Segment.—This segment shows a similar picture as seen in the eighth thoracic. The column of Goll on the right side is much more degenerated than that on the left. The columns of Burdach are not involved. The same is true of the root entrance zone and cornu-commissural zone of each side.

With the hemalum and acid-fuchsin stain all the segments show the same picture. The pia arachnoid shows a slight thickening with slight round cell infiltration.

With the Nissl stain the cells of the anterior and posterior horns are found considerably pigmented.

Pathological Summary.—The degeneration of the posterior root zone was pronounced through all the sacral segments, and extended as high as the fourth lumbar segment. It was slight at the third lumbar segment, and in the middle thoracic and lower cervical segments only the columns of Goll were involved. The degeneration was throughout more pronounced on the right side and involved the posterior root zone from the fourth lumbar down on the right, whereas on the left side in the corresponding area many normal root fibers entered the posterior horns.

In this case preservation of the patellar tendon reflexes and the disappearance of the Achilles tendon reflexes are satisfactorily explained by the pathological findings. Ordinarily in tabes the lumbar segments are involved, and the typical symptoms arise, including loss of patellar reflex. In this case the sacral segments showed pronounced degeneration, the lower lumbar segments were involved, but the pathway for the reflex arc had not been destroyed. The slight involvement of a few of the fibers of the reflex collaterals of the right side corresponds to the finding of a slight diminution of the right patellar tendon reflex.

Achard and Levi, in 1897, reported a similar case, in which the diagnosis of tabes was made during life. The persistence of the reflexes was the chief feature. The patient had the Argyll-Robertson pupils, tabetic gait, and Romberg sign. The reflexes were examined at different times and showed no diminution or exaggeration. The microscopical examination showed, as in my case, extensive sclerosis in the sacral segments, especially in the inferior, while the upper sacral and all the lumbar segments showed a limited area of sclerosis. T. Pineles describes a case of sacral tabes in a patient aged 46 years, in which besides lancinating pains and Argyll-Robertson pupils, the left patellar tendon reflex was lost, while the right patellar tendon reflex was obtainable only by reinforcement. There was no ataxia or Romberg. He speaks of it as a case of incipient tabes, in which the disease began in the lower spinal segments.

Redlich[2] describes in his monograph on tabes a case of tabes dementia paralytica in which the preservation of the patellar tendon reflexes was noted, together with loss of Achilles tendon reflex. The tabetic changes were found in the lower lumbar and sacral segments. In the upper sacral and lower lumbar segments the picture was that of an incipient process, while the lower sacral segments showed scarcely any changes. In my case an incipient process was not to be considered in as much as the degeneration throughout the sacral segments was so extensive.

In Pick's[3] case there was left facial paralysis, and the left patellar reflex was absent, while the right was only obtainable by Jendrassik's method. Sections showed intactness of the reflex arc in the lumbar segments.

Babinski and Nageotte, in 1905, reported their findings in a case, but according to the findings it was not one of the sacral type.

The unique process in their case I will discuss later; let us now return to the question of the patellar tendon reflex as an early symptom. What emphasis should be laid upon this symptom? Westphal in his first study of the knee-jerk said, "it is present when the posterior columns are involved only as far as the lower thoracic and upper lumbar segments, and also when the lateral pyramidal tracts are involved." Clinical cases have been reported by Hamilton, Erb, Theme, Berger and others, in which the patellar tendon reflexes were preserved. Berger found it preserved in 2.4 per cent. of 82 cases examined. Abadie found in 196 cases 13.5 per cent. with preservation of the patellar tendon reflexes. The studies of Jackson and Taylor, Goldflam, Mamlock, Dercum and others in cases of tabes with secondary hemiplegia and return of the patellar tendon reflex serve to show us the variation of this reflex.

To-day more stress is laid upon the disappearance of the

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[2] Redlich, E. "Die Pathologie der tabischen Hinterstrangserkrankung," 1897.

[3] Pick. "Anatomische Befund bei einseitigen Fehlen des Knie phaenomens." Arch. f. Psych., Bd. xx.

Achilles tendon reflex as an earlier symptom. Bregman quotes Babinski and Goldflam's view that the Achilles tendon reflex is more frequently and earlier altered than is the patellar reflex. The latest studies of collected cases of tabes seem to indicate the correctness of this statement. Kollarits, a Hungarian observer, found the Achilles tendon-jerk absent in 65 of 100 cases examined, while the patellar tendon reflexes were absent in 56, thus making 11 cases with absent Achilles tendon reflex as the only sign. Sarbo analyzed 92 cases and found the patellar tendon reflex abnormal in 87 per cent., the Achilles tendon reflex abnormal in 91 per cent. Bregman asserts from these, together with the sensory findings, that the sacral portion of the spinal cord is the earliest portion affected in tabes.

How shall we account pathologically for these variations in the patellar tendon reflex? We have seen that tabes may occur in which the patellar tendon reflex is preserved; again, we see cases in which the reflex disappears only to reappear. The view of most observers to-day is that the integrity of the patellar and Achilles tendon reflex depends upon two factors: first and chiefly, the preservation of the fibers forming the reflex arc, and secondly, upon a cortical influence whose pathway may be in the pyramidal tract, or in the cerebellar tract. If the reflex arc is entirely destroyed, then the patellar tendon reflex is entirely lost, and this is the picture described in most cases of tabes, pathologically the most pronounced degeneration is seen in the lumbar segments. If these segments are only partially destroyed then the patellar tendon reflex may or may not be present, or it may be absent and return. This phenomenon was seen in those hemiplegic tabetics, and depends upon the removal of cortical inhibition by degeneration of the pyramidal tracts.

In Pick's case where there had been return of the knee-jerks, the pathological findings explained the symptoms, in that only a portion of the reflex arc had been destroyed, and with the secondary advent of the destruction of the pyramidal tract the removal of inhibition occurred upon the few fibers of the reflex arc left intact. In Achard's case, in Redlich's case and in my case, the preservation of the reflex was explained by the involvement only of the sacral and lower lumbar portions of the cord.

Babinski and Nageotte[4] found an inverse systematization in their case. A young man, 25 years of age, had tabetic symptoms, with preservation of the patellar tendon reflex. On examination of the sections the degeneration involved the long fibers of the posterior roots, or in other words, the postero-external zone. The oval field of Flechsig and the columns of Goll presented lesions of the same intensity while the short or medium systems of fibers, those usually attacked first in this disease, escaped. This is contrary to the usual onset of this disease, consequently they speak of it as an inverse systematization in tabes. No mention, however, is made of the sacral segments. The important observation to be deduced from my case, and those cited is that, clinically, tabes should not be overlooked in those cases in which the patellar tendon reflex is preserved. The early disappearance of the tendo-Achillis reflex is attaining greater significance, and as indicated above, among some observers, as Babinski and Bregman, is of greater value as an early sign.

Besides this case also shows the polymorphic nature of this disease, and to-day instead of only the lumbar type, the cervical type, the optic type, and paralytic type, we have pathologically and clinically the sacral type.

I wish to take this opportunity to thank Dr. Spiller for his kind assistance in the examination of the pathological sections.

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[4] Babinski and Nageotte. "Note sur un cas de tabes a systematization exceptionelle." Soc. de Biol. de Paris, 1905.